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Anomalous Presentation and Endoscopic Findings of Acute Esophageal Necrosis: A Case Report

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Anomalous Presentation and Endoscopic Findings of Acute Esophageal Necrosis: A Case Report

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Abstract

Introduction:

Acute esophageal necrosis (AEN), is a rare disorder with an estimated incidence of 0.01-0.28%. It typically affects the distal third of the esophagus. Approximately 90% of patients with acute esophageal necrosis present with upper gastrointestinal (GI) bleeding, primarily due to ischemic insult or severe hemodynamic compromise. In this report, we describe a case in which the patient presented with odynophagia and dysphagia, but without upper GI bleeding. The patient exhibited pan mucosal involvement of the esophagus, which extended to the gastric antrum.

Case: A 55-year-old man with a history of type 2 diabetes mellitus and substance abuse presented with intractable nausea and vomiting, leading to hospitalization for hypovolemic shock secondary to diabetic ketoacidosis (DKA). Following the resolution of DKA, he experienced worsening dysphagia and odynophagia. A barium swallow of the esophagus revealed a distal esophageal stricture. Upper endoscopy revealed marked hyperpigmentation, extensive sloughing, and mucosal friability involving the entire esophageal mucosa. Diffuse erythema, sloughing, and mucosal friability extended from the fundus to the antrum of the stomach, accompanied by mild pyloric stenosis and food retention. The patient was diagnosed with acute esophageal necrosis and marked acid reflux due to gastric hypomotility.

Treatment included intravenous proton pump inhibitors and total parenteral nutrition for two weeks. A follow-up upper endoscopy showed healing esophagitis and duodenitis, with the presence of moderate-sized clean based duodenal ulcers. The patient was maintained on a high dose of pantoprazole for a total of eight weeks.

Discussion: In contrast to the typical presentation of upper gastrointestinal (GI) bleeding in acute esophageal necrosis (AEN), our patient exhibited symptoms of vomiting, dysphagia, and odynophagia. Endoscopically, we observed diffuse pan esophageal necrosis and diffuse involvement of the gastric mucosa, which differed from the expected restriction to the distal esophagus with a distinct demarcation at the gastroesophageal junction (GEJ). Consequently, our case emphasizes the importance of considering AEN as a potential diagnosis in critically ill patients who present with complaints of dysphagia and odynophagia, even in the absence of upper GI bleeding. The atypical presentation and endoscopic findings observed in our patient warrant further investigation through additional studies to enhance our understanding of AEN.

Keywords

Black Esophagus, Acute Esophageal Necrosis, Endoscopy, Gastroenterology, Esophagus, Necrosis

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Conflict of Interest Statement

The authors of this case report declare that there are no conflicts of interest to disclose regarding the

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CASE REPORT

Anomalous Presentation and Endoscopic Findings of Acute Esophageal Necrosis: A Case Report

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Abstract

Introduction: **Acute esophageal necrosis (AEN), is a rare disorder with an estimated incidence of 0.01−0.28%. It typically affects the distal third of the esophagus. Approximately 90% of patients with acute esophageal necrosis present with upper gastrointestinal (GI) bleeding, primarily due to ischemic insult or severe hemodynamic compromise. In this report, we describe a case in which the patient presented with odynophagia and dysphagia, but without upper GI bleeding. The patient exhibited pan mucosal involvement of the esophagus, which extended to the gastric antrum.**

Case: **A 55-year-old man with a history of type 2 diabetes mellitus and substance abuse presented with intractable nausea and vomiting, leading to hospitalization for hypovolemic shock secondary to diabetic ketoacidosis (DKA). Following the resolution of DKA, he experienced worsening dysphagia and odynophagia. A barium swallow of the esophagus revealed a distal esophageal stricture. Upper endoscopy revealed marked hyperpigmentation, extensive sloughing, and mucosal friability involving the entire esophageal mucosa. Diffuse erythema, sloughing, and mucosal friability extended from the fundus to the antrum of the stomach, accompanied by mild pyloric stenosis and food retention. The patient was diagnosed with acute esophageal necrosis and marked acid reflux due to gastric hypomotility.**

Treatment included intravenous proton pump inhibitors and total parenteral nutrition for two weeks. A follow-up upper endoscopy showed healing esophagitis and duodenitis, with the presence of moderate-sized clean based duodenal ulcers. The patient was maintained on a high dose of pantoprazole for a total of eight weeks.

Discussion: **In contrast to the typical presentation of upper gastrointestinal (GI) bleeding in acute esophageal necrosis (AEN), our patient exhibited symptoms of vomiting, dysphagia, and odynophagia. Endoscopically, we observed diffuse pan esophageal necrosis and diffuse involvement of the gastric mucosa, which differed from the expected restriction to the distal esophagus with a distinct demarcation at the gastroesophageal junction (GEJ). Consequently, our case emphasizes the importance of considering AEN as a potential diagnosis in critically ill patients who present with complaints of dysphagia and odynophagia, even in the absence of upper GI bleeding. The atypical presentation and endoscopic findings observed in our patient warrant further investigation through additional studies to enhance our understanding of AEN.**

Keywords: Black esophagus, Acute esophageal necrosis, Endoscopy, Gastroenterology, Esophagus, Necrosis

1. Introduction

Acute esophageal necrosis (AEN), also known as black esophagus, is a rare clinical entity characterized by circumferential ischemia of the distal esophagus in the setting of ischemia. In 97% of cases, necrosis manifests in the distal segment of the esophagus.[1] It has a prevalence ranging from 0.0001% to 0.28% on esophagogastroduodenoscopies. Males are four times more likely to be affected than females, with a higher prevalence observed in older males in their sixth decade of life.[2–6]

The endoscopic findings reveal mucosal necrosis of the distal segment of the esophagus that extends from the gastroesophageal junction proximally. It is

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atypical for the necrosis to encompass the whole esophagus or to extend beyond the gastroesophageal junction. The black esophageal mucosa is likely due to a vascular watershed area and a reduced vascular supply in the distal segment of the esophagus.[2,4] The mortality rate ranges between 32% and 38%, depending on the severity of the underlying precipitating causes. Specifically, 6% of deaths can be directly attributed to AEN.[4,5,7]

We present a unique and atypical presentation of AEN, where our patient had necrosis involving the entire esophagus and extending beyond the gastroesophageal junction with diffuse gastric mucosal involvement.

2. Case discussion

A 55-year-old man with a past medical history of insulin-dependent type 2 diabetes mellitus, substance abuse (cannabis and cocaine), asthma, and peripheral neuropathy presented with intractable nausea and vomiting. His initial vitals showed a blood pressure of 197/93 mmhg, heart rate of 110 beats per minute, temperature of 95.8 F, and respiratory rate of 24 breaths per minute.

The patient had been non-compliant with his home insulin regimen and complained of generalized abdominal pain, which had worsened gradually. He also experienced associated anorexia and poor oral intake. His lab work revealed a white blood cell count of 22,600/mm^3, hemoglobin of 20.5 g/dl, sodium of 127 mEq/L, potassium of 4.4 mEq/L, bicarbonate of less than 10 mEq/L, blood glucose of 493 mg/dl, B-hydroxybutyrate of 6.66 mmol/L, lactic acid of 2.1 mmoL/l and an incalculable anion gap. Urinalysis was positive for ketones. Subsequently, he was admitted to the medical intensive care unit for further management of diabetic ketoacidosis (DKA).

Following the resolution of DKA, he experienced worsening dysphagia and odynophagia, along with complaints of regurgitation. The gastroenterology team was consulted, who initially recommended a barium swallow of the esophagus which revealed a distal esophageal stricture. Subsequent upper endoscopy revealed marked hyperpigmentation, extensive sloughing, and mucosal friability involving the entire esophageal mucosa (Fig. 1). These findings extended from the fundus to the antrum of the stomach, accompanied by mild pyloric stenosis and food retention consistent with gastroparesis. The patient was diagnosed with acute esophageal necrosis and marked acid reflux due to gastric hypomotility. The patient was kept nil per os and started on intravenous proton pump inhibitors along with total parenteral nutrition for two weeks.

The patient underwent a follow-up upper endoscopy 20 days later, revealing healing esophagitis and duodenitis, along with moderate-sized clean-base duodenal ulcers (Fig. 2). The patient continued to receive pantoprazole 40 mg twice daily for a total of eight weeks and demonstrated significant improvement in his symptoms.

3. Discussion

Acute esophageal necrosis (AEN), also known as Gurvits syndrome, black esophagus, and necrotizing esophagitis, is a rare disease. It is characterized by a diffuse circumferential black appearance of the esophageal mucosa, affecting the distal esophagus and ending distinctly at the gastroesophageal junction.[2] Black esophagus was first described in 1990 by Goldenberg et al. in a postoperative patient requiring esophagectomy for esophageal ischemic stricture.[5,8] It was presumed to be a rare entity with an estimated prevalence of 0.2% in autopsy series. However, the actual

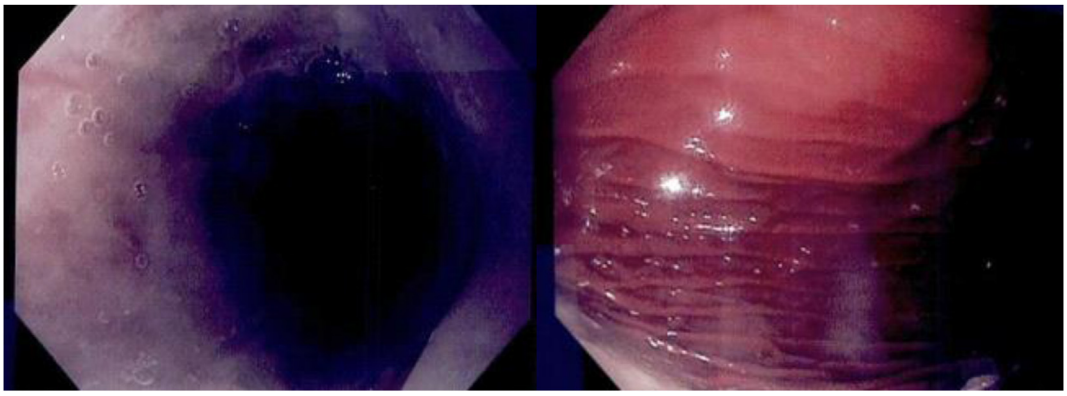

Fig. 1. Initial endoscopy showing AEN.

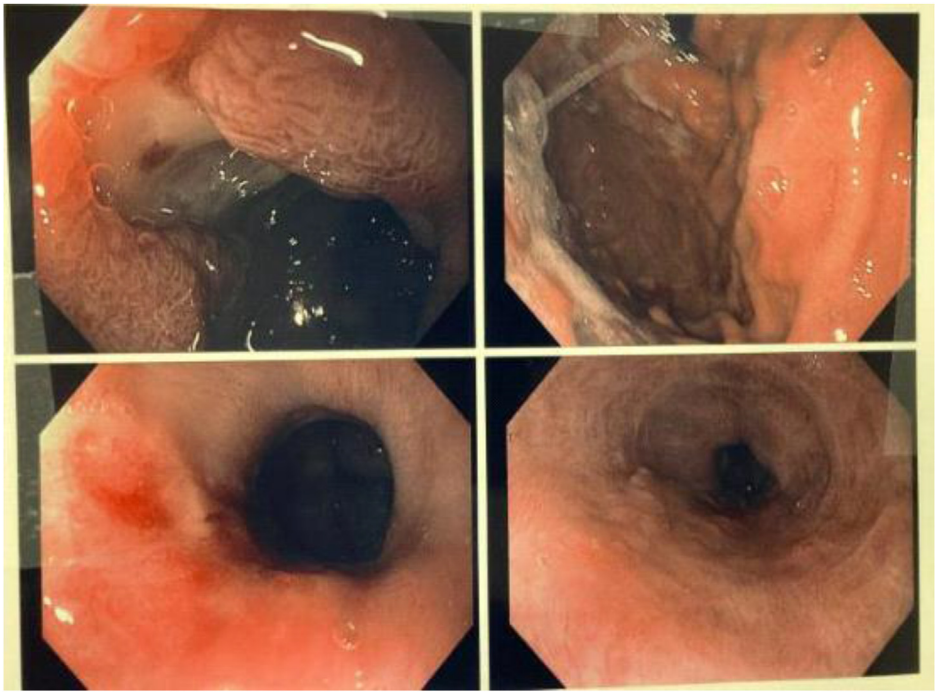

Fig. 2. Follow-up endoscopy 20 days later showing AEN resolution.

prevalence could be higher, possibly due to the esophageal mucosa's propensity to heal and the misleading clinical picture.[3] AEN has become more commonly recognized and described due to the periodic practice of performing endoscopy.[5]

The typical clinical presentation is hematemesis, melena, and anemia in 90% of cases. Other symptoms include abdominal pain, fever, dysphagia, nausea, and syncope.[3,4,6] Differential diagnoses can encompass malignant melanoma, acanthosis nigricans, corrosive agents ingestion, dye ingestion, dissecans superficialis, intraepithelial hemorrhage of the esophagus, pseudo melanosis, and infectious diseases.[3,5,6,9,10] In our case, the patient presented with odynophagia and dysphagia but without upper GI bleeding. The etiology of AEN is complex; predisposing factors include tissue ischemia observed in cardiovascular compromise, decreased mucosal defenses in the esophagus caused by malignancies and malnutrition, reflux of acidic contents of the stomach, and obstruction at the gastro-duodenal junction. In addition, duodenal ulcers, uncontrolled diabetes, chronic liver disease, chronic kidney disease, hypertension, shock, antibiotics, and alcohol abuse, have been associated with AEN.[2–4]

Several studies support the two-hit hypothesis, which involves initial tissue hypoperfusion combined with reduced mucosal defense, predisposing to insult from the acidic gastric contents.[5] The distal third of the esophagus has reduced vascular supply compared to the proximal segment. It is speculated that the chain of events starts with hypovolemia, which triggers acid reflux. Obstruction at the gastric outlet further aggravates the situation by pooling the acid contents in the stomach, leading to further ongoing reflux that damages the esophagus, ultimately progressing to necrosis.

This theory finds support in observations where curtailing vascular supply results in necrosis, which significantly improves upon the restoration of blood flow. Interestingly, our patient did not experience hypotensive episodes and did not require to be started on vasopressors while admitted to the medical intensive care unit.[2] The current literature supports the ischemic etiology hypothesis, noting that the esophageal mucosal necrosis predominates in the distal third of the esophagus which has reduced vascularization.[4,11]

The endoscopic findings reveal mucosal necrosis of the distal segment of the esophagus, extending proximally. Typically, the necrosis doesn't continue beyond the gastroesophageal junction.[2] Several studies have reported that the esophageal necrosis doesn't extend beyond and discernibly ends at the gastroesophageal junction. Similarly, endoscopy confirms the diagnosis of AEN by revealing diffuse circumferential discoloration of the distal esophagus which ends at the Z-line.[3,5,11] In terms of the extent of proximal involvement, a systematic review of 160 cases reported that in 64.3% of the cases, necrosis

extended up to the middle third of the esophagus and 92.9% were localized in the distal third. The necrosis abruptly ends at the Z-line.[12] In contrast, our patient had diffuse esophageal necrosis and diffuse involvement of the gastric mucosa. Upper endoscopy revealed marked hyperpigmentation, extensive sloughing, and mucosal friability. Diffuse erythema, sloughing, and mucosal friability extended from the fundus to the antrum of the stomach, which differed from the expected restriction to the distal esophagus with a distinct demarcation at the gastroesophageal junction (GEJ). This is an atypical presentation as it extends beyond the Z-line, standing against the watershed theory mentioned above.

AEN can be classified according to the stage as seen on endoscopy.[1] Stage 0 designates a pre-necrotic viable esophagus, while stage 1 refers to an acutely diseased organ. The endoscopic picture is dominated by a striking diffuse, circumferential, black-appearing esophageal mucosa with occasional yellow exudates and signs of friability, loss of light reflex, rigidity, and under-distension of the lumen. These endoscopic findings nearly universally start at the gastroesophageal junction, involve the distal esophagus, and variably extend proximally. The histological appearance is notable for the lack of viable squamous epithelium, and the pronounced mucosal necrosis and necrotic debris with possible extension into the submucosa and muscularis propria.[1] Associated findings may include heavy leukocytic infiltration with severe inflammatory changes, visible vascular thrombi, deranged muscle fibers, and mucosal infection with viral, fungal, and bacterial pathogens. Stage 2 describes the healing phase of AEN dominated by residual black areas in the esophagus and thick white exudates composed of necrotic debris that cover friable pink mucosa. This “chessboard” appearance can sometimes be the presenting endoscopic picture of AEN on delayed endoscopy. The exact timing of this change is unknown, and likely parallels the underlying general condition of the patient. However, it has been observed as late as one month after the diagnosis. Stage 2 is also notable for some improvement in the histological appearance of the esophageal tissue, with scattered areas of necrosis among underlying cellular regeneration, granulation tissue, and inflammatory changes. As early as 1−2 weeks after diagnosis, the esophageal mucosa acquires its normal endoscopic appearance in stage 3 of the AEN with only microscopically present granulation tissue, a sequela of recent injury. Endoscopy is usually conclusive; histopathology can be supportive for confirmation, identifying necrotic tissue, loss of epithelium, granulation tissue with leukocytic infiltrates extending into the submucosa, and muscularis propria.[1,4,6,7]

Management is directed towards rectifying the compromise in blood flow and reversing the factors causing the patient's hemodynamic instability. In most cases, restricting oral diet and intravenous PPI can often reverse the tissue injury. Nasogastric tube insertion is not advisable due to the significant risk of perforation. Antibiotics are recommended for patients with suspected esophageal perforation, sepsis, or immunocompromised states.[4] Treating the underlying medical condition is imperative. Managing concomitant comorbidities is essential to prevent disease aggravation, ensuring a favorable prognosis and decreasing mortality rates.[4,9] Complications may develop in some cases, such as esophageal stenosis and strictures, which can be treated with endoscopic dilatation.[2] Perforation and resultant mediastinitis, tracheoesophageal fistula, and abscess formation can also occur potentially requiring surgical interventions.[3,9] The mortality rate ranges between 32% and 38%, depending on the severity of the underlying precipitating causes and the age of the patient. Additional research is required to explore the elevated occurrence of AEN in males, particularly in the elderly, as the literature review did not provide information elucidating the potential causes. Death usually results from the underlying condition, with only 6% of the deaths directly attributed to AEN.[4,5,7,13]

Our case emphasizes the importance of considering AEN as a potential differential diagnosis in critically ill patients who present with complaints of dysphagia and odynophagia, even in the absence of upper GI bleeding. The atypical presentation and endoscopic findings observed in our patient warrant further research and studies to enhance our understanding of AEN.

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Conflicts of interest

None declared.

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